

1 1. A substantially pure mannin-binding lectin
2 associated serine protease-2 (MASP-2) polypeptide.

1 2. The polypeptide of claim 1, said polypeptide
2 being capable of associating with mannan-binding lectin
3 (MBL).

1 3. The polypeptide of claim 1, said polypeptide
2 being conjugated to a label or toxin.

1 4. A polypeptide containing the sequence
2 identified as SEQ ID NO. 1.

1 5. A polypeptide according to claim 4 with a
2 molecular mass of 20K.

1 6. A polypeptide with a molecular mass of 52K and
2 containing the sequence identified as SEQ ID NO 1.

1 7. The polypeptide of claim 1, said polypeptide
2 having serine protease activity.

1 8. A polypeptide of claim 1, said polypeptide being
2 capable of MASP-2 activity in an *in vitro* assay for MBLectin
3 complement pathway function.

1 9. A polypeptide according to claim 1, said
2 polypeptide being capable of competitively inhibiting MASP-2
3 serine protease activity.

1 10. A polypeptide according to claim 1 comprising a
2 fragment of the polypeptide of SEQ ID NO:2, said polypeptide
3 being a competitive inhibitor of complexing of MBL/MASP-2.

1 11. A polypeptide according to claim 5 or claim 6,
2 said polypeptide having the amino acid sequence of SEQ ID
3 NO:2.

1 12. A compound capable of competitively inhibiting
2 serine protease activity of MASP-2 or a fragment thereof.

1 13. An isolated nucleic acid molecule of claim, the
2 molecule comprising a nucleotide sequence encoding a
3 polypeptide having sequence that is at least 85% identical
4 to the sequence of SEQ ID NO:1 or 2.

1 14. An isolated nucleic acid sequence encoding a
2 mannan-binding lectin associated serine protease-2 (MASP-2)
3 polypeptide according to claim 1.

1 15. A nucleic acid vector comprising the nucleic
2 acid molecule of claim 14.

1 16. The nucleic acid vector of claim 15 wherein
2 said vector is an expression vector.

1 17. The vector of claim 16, further comprising a
2 regulatory element.

1 18. An antibody produced by administering an MASP-2
2 polypeptide according to claim 1 to an antibody producing
3 animal.

1 19. An antibody that selectively binds to MASP-2.

1 20. The antibody of claim 18 or claim 19, wherein
2 said antibody is a monoclonal antibody.

1 21. The antibody of claim 18 or 19, said antibody
2 being coupled to a compound comprising a detectable marker.

1 22. A pharmaceutical composition comprising the
2 polypeptide of claim 1 or the antibody of claims 18 or 19.

1 23. A method for detecting mannin-binding lectin
2 associated serine protease-2 (MASP-2), said method
3 comprising:

4 (a) obtaining a biological sample;

5 (b) contacting said biological sample with a MASP-2
6 polypeptide specific binding partner that specifically binds
7 MASP-2; and

8 (c) detecting said complexes, if any, as an
9 indication of the presence of mannin-binding lectin
10 associated serine protease-2 in said sample.

1 24. A method according to claim 23, in which the
2 specific binding partner is an antibody.

1 25. A method for detecting MASP-2, said method
2 comprising an assay for MASP-2 complement fixing activity.

1 26. The methods of claims 23 or 24 for quantitative
2 assay of MASP-2 or MASP-2 activity in biological samples.

1 27. A method for detecting MASP-2 nucleic acid
2 expression, comprising detecting RNA having a sequence
3 encoding a MASP-2 polypeptide by mixing the sample with a
4 nucleic acid probe that specifically hybridizes under
5 stringent conditions to the nucleic acid of claim 13 or 14.

1 28. A method for treating patients deficient in
2 MASP-2 by administering to the patient the peptide of claim
3 1.

1 29. A method for treating patients deficient in
2 MASP-2 by administering to the patient nucleic acid
3 according to claim 13 or 14.

1 30. A method for inhibiting the activity of MASP-2
2 by administering to the subject a compound that inhibits
3 expression or activity of MASP-2.

1 31. The method of claim 27 in which the compound is
2 a MASP-2 anti-sense nucleic acid sequence.

1 32. The method of claim 30 comprising administering
2 a compound that inhibits complexing of MBL and MASP-2.

1 33. An assay for polymorphisms in the nucleic acid
2 sequence encoding MASP-2.

1 34. A method of detecting the presence of MASP-2-
2 encoding nucleic acid in a sample, comprising mixing the
3 sample with at least one nucleic acid probe capable of
4 forming a complex with MASP-2-encoding nucleic acid under
5 stringent conditions, and determining whether the probe is
6 bound to sample nucleic acid.

1 35. A nucleic acid probe capable of forming a
2 complex with MASP-2-encoding nucleic acid under stringent
3 conditions.

1 36. An assay for polymorphisms in the polypeptide
2 sequence comprising MASP-2 or its precursor.

1 37. A method for diagnosing a disorder associated
2 with aberrant expression of MASP-2, comprising obtaining a
3 biological sample from a patient and measuring MASP-2
4 expression in said biological sample, wherein increased or
5 decreased MASP-2 expression in said biological sample
6 compared to a control indicates that said patient suffers
7 from a disorder associated with aberrant expression of MASP-
8 2.

1 38. A method for diagnosing a disorder associated
2 with aberrant activity of MASP-2, comprising obtaining a
3 biological sample from a patient and measuring MASP-2
4 activity in said biological sample, wherein increased or
5 decreased MASP-2 activity in said biological sample compared
6 to a control indicates that said patient suffers from a
7 disorder associated with aberrant activity of MASP-2.

1 39. A method of assaying for activity MBL-complexed
2 MASP, the method comprising
3 providing a sample to be assayed and substantially
4 reducing any artifact resulting from activation of the
5 classical complement fixing pathway by conducting the assay
6 in the presence of an ionic strength high enough to
7 effectively reduce activation of the classical complement
8 fixing pathway but not so high as to substantially interfere
9 with activity of MBL-complexed MASP.